Strudel Hematology Review

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Primary Hemostasis

1) Endothelial injury
2) vWF binds to exposed subendothelial collagen (Produced from WP bodies of endothelial cells, alpha granules of plt)
3) Plt binds vWF (GP1B)->induces conformational change
4) Plt releases ADP, **Ca2+**, TXA2
5) ADP binds P2Y12 to induce gpIIB/IIIa expression on plt
6) Fibrinogen binds gpIIb/IIIa and provides weak platelet plug ("white thrombus")

Factors favoring aggregation?

- TXA2
- Reduced blood flow/stasis

Factors preventing aggregation?

- PGI2, NO
- Increased blood flow

What stabilizes the platelet plug?

Secondary hemostasis – the fibrin "red" clot

Quick review

MOA of antiplatelet agents:

Aspirin (ASA)?
Inhibits COX 1 and COX2
Irreversible
Prevents synthesis of TXA2 -> less platelet activation, less vasoconstriction

Clopidogrel (Plavix), prasugrel, ticlopidine?
Irreversibly binds P2Y12 ADP receptors on plt.
Prevents activation of the glycoprotein GPIIb/IIIa complex

Abciximab, eptifibatide, tirofiban?
Inhibit GPIIb/IIIa

The Coagulation Cascade

Intrinsic

XII(a)->XI(a)->IX(a)->VIII(a)->common pathway X(a)->V(a)->II(a)->I(a)

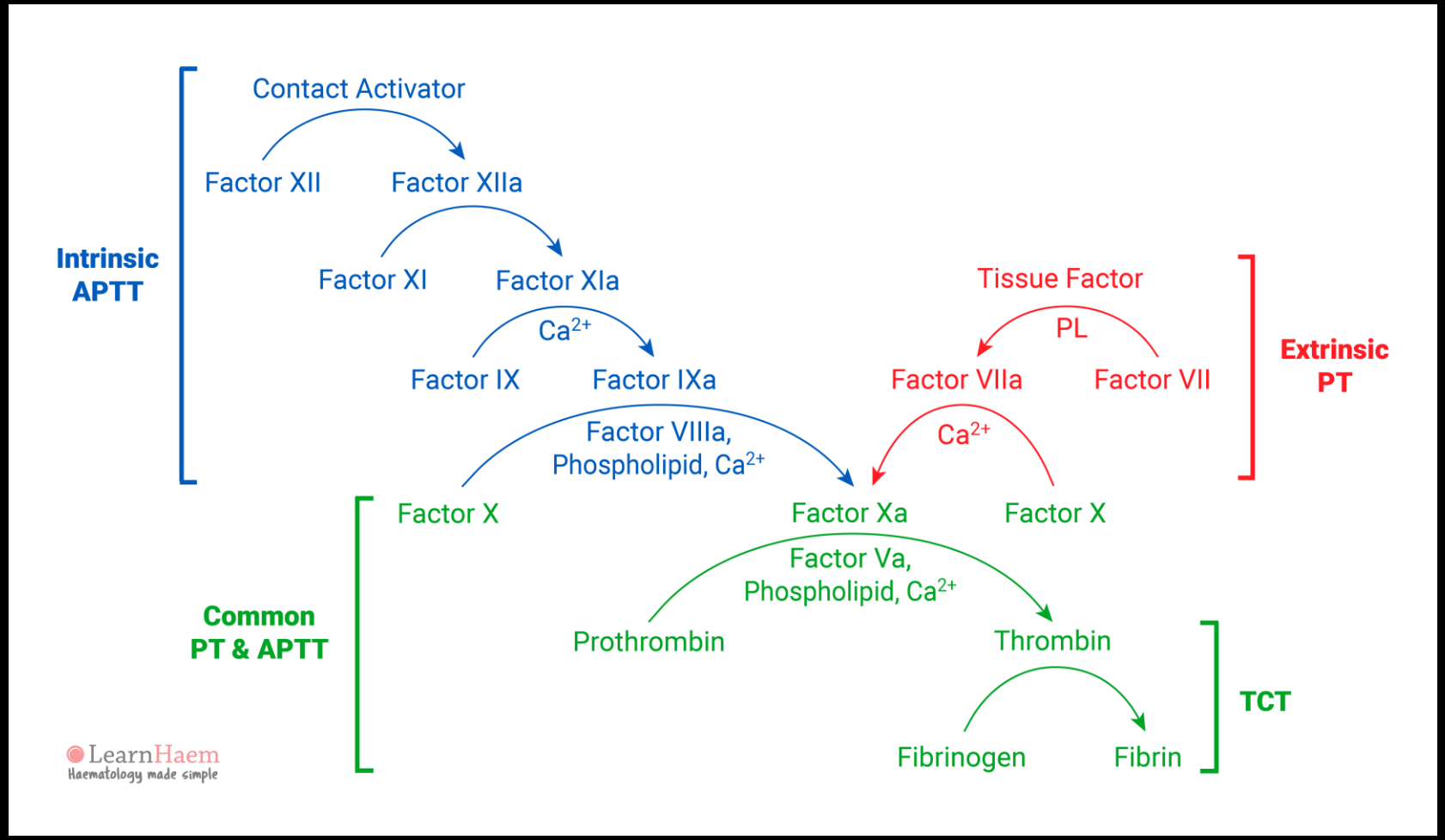
IIa also activates XIII

How to remember?
TENET

How to measure?
aPTT

Extrinsic:
VII(a)->X(a) -> common pathway X(a)->V(a)->II(a)->I(a)

How to measure?
PT



https://www.learnhaem.com/courses/coagulation/lessons/normal-haemostasis/topic/the-revised-coagulation-cascade/

Anticoagulant review

Warfarin (coumadin) MOA?
Epoxide reductase inhibitor (effectively inhibits Vitamin K-mediated gamma-carboxylation of factors – required for activation)
Originally used as rat poison (blood thinner)
What factors are affected?
II, VII, IX, X, Protein C, S
Requires routine INR measurement/titration
Reversal?
Vit K
Rapid reversal?
FFP, PCC
Bridge with?
Heparin/Heparin derivatives
Which factor has shortest half life?
VII

Reason for bridging?
Warfarin-induced skin necrosis
Etiology?
Protein C has shorter half life -> paradoxical hypercoagulable state

Wisconsin Alumni Research Foundation

Warfarin Scenarios

70 y.o. M on warfarin is diagnosed with a hemorrhagic stroke. Management?

Hold warfarin, reverse emergently with Vit K + PCC

50 y.o. F on warfarin is found to have an INR of 6, but is not displaying any signs of bleeding. Management?

Hold warfarin, recheck INR ~48 hrs later

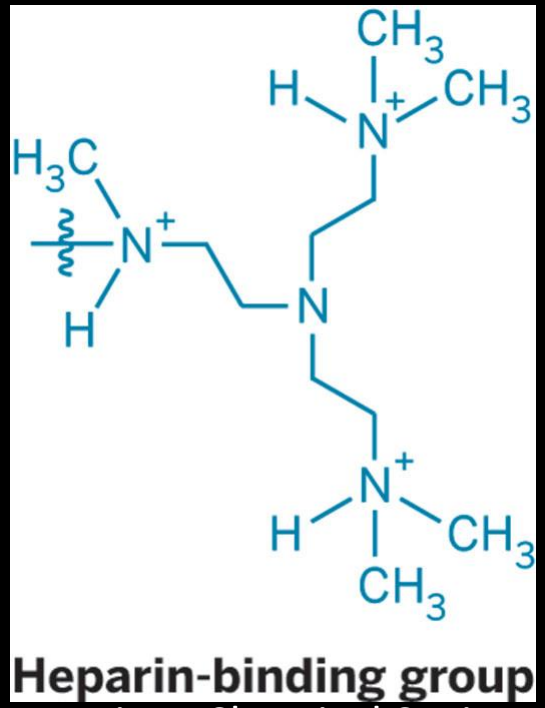
60 y.o. M on warfarin has an INR of 12, and is not displaying any signs of bleeding. Management?

Hold warfarin, give a dose of vit K, recheck INR

Anticoagulant review

Heparin?
Enhances ATIII -> inhibits II, VIIa, IXa, Xa, XIa, XIIa
Short half life, can be given IV as drip. Monitor w/ PTT
Clearance?
Hepatic
Reversal?
Protamine sulfate (pos charge) - from salmon sperm protein

Dalteparin, enoxaparin (lovenox)?
LWMH, Act more on Xa. Can be given subq.
Clearance?
Renal
Reversal?
Still protamine – not as effective



American Chemical Society


GoodRx

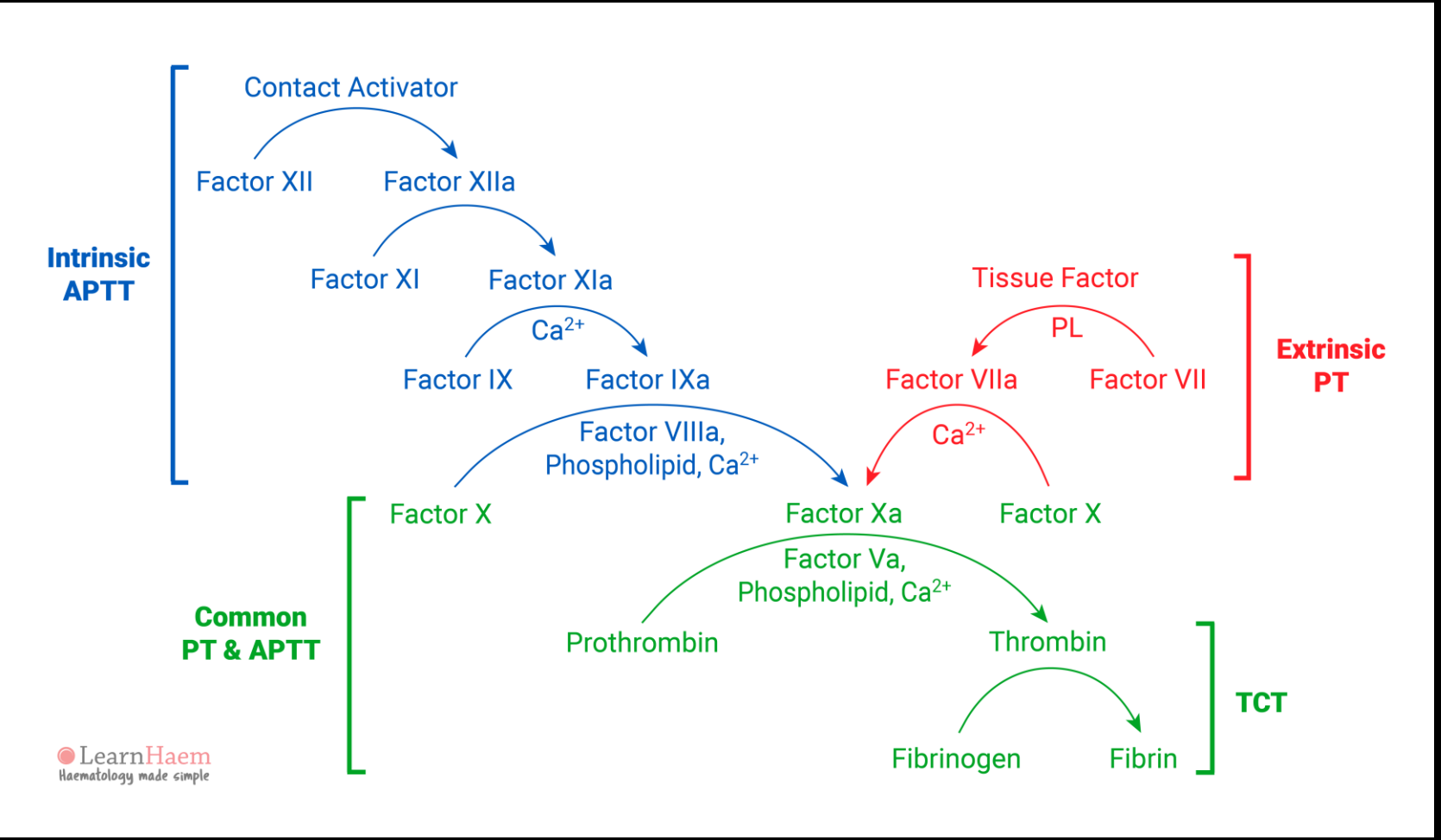
Anticoagulants (continued)

Adverse effect of heparin/derivatives?

Heparin induced thrombocytopenia (HIT)
More likely with unfractionated heparin but may occur w/ LWMH.
2 subtypes:

Type 1?
"Non-immune heparin-associated TCP" - direct platelet aggregation, within 5 days, mild
Next best step?
Observation
Type 2?
"Immune associated" characterized by formation of IgG ab against plt (heparin PF4)
Next best step?
d/c heparin immediately, switch to bivalirudin/argatroban (direct thrombin inhibitors)
How to dx?
Used to be serotonin release assay, now is a PF4 serology

Clotting



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3 yo M presents to ED after ground level fall onto knees 1 hour ago. Now with bilateral knee pain/swelling with hematomas on exam and hemarthrosis on arthrocentesis. Family history of easy bleeding and bruising. Dx?

Heme

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Hemophilia

Most common?

Hemophilia A (VIII)

Pathophys?

Impaired secondary hemostasis. Hemophilia A = factor VIII; hemophilia B = factor IX def; hemophilia C = factor XI def

Clinical presentation?

Easy bruising/bleeding, hematomas, prolonged bleeding, oozing after tooth extraction, GI bleeding, hematuria, epistaxis, hemarthrosis, brain bleed

Labs?

PTT prolonged with normal PT; Bleed time and PLTs nml

Does it correct with mixing?

YES

Treatment?

Severe is treated with recombinant or plasma derived factor VIII or IX.

Mild hemophilia A can be treated with desmopressin - increases vWF release which stabilizes factor VIII

How do you distinguish acquired from genetic/congenital?

Acquired appears later in life d/t autoantibody formation against the replaced factor ("acquired inhibitor")

Mixing study will not correct now

BONUS: What would you see with repeated hemarthroses?

Joint breakdown due to hemosiderin deposition over time

25 year old woman presents with 2 days of a red, swollen calf. Has not had similar episode in the past. Denies shortness of breath. On exam, you appreciate tenderness to palpation of RLE and pain with dorsiflexion of right foot. What is your next step in management and likely dx?

Heme

25 year old woman presents with 2 days of a red, swollen calf. Has not had similar episode in the past. Denies shortness of breath. On exam, you appreciate tenderness to palpation of RLE and pain with dorsiflexion of right foot. What is your next step in management and likely dx?
U/S, DVT/VTE

What genetic predisposition may you suspect?
Factor V leiden (most common), prothrombin 20210a mutation
Protein C, Protein S deficiency, Antithrombin deficiency

Would you obtain genetic testing at this time?
No, as this is the first instance of VTE and is not life threatening VTE-may consider testing if multiple unexplained CTE or arterial thrombosis. Would not affect management in this instance.

What are other causes of hypercoagulability?
Antiphospholipid syndrome (APS)

How would this present in test questions?
Recurrent pregnancy loss due to placental thrombosis

Tx?
LWMH

38 yo previously healthy F presents w/ epistaxis x 1 day. + IVDU. There is blood in the nares and petechiae distal to the blood pressure cuff. PLTs 28,000.

More Platelets

38 yo previously healthy F presents w/ epistaxis x 1 day. + IVDU. There is blood in the nares and petechiae distal to the blood pressure cuff. PLTs 28,000.
Immune Thrombocytopenia (ITP)

Pathophysiology?
Acquired immune-mediated destruction of platelets. IgG antibodies directed against platelet membrane glycoproteins such as GPIIb/IIIa (primary has idiopathic cause, secondary causes can be triggered by cancer, infection, or other autoimmune conditions – antiphospholipid syndrome or lupus)

Clinical presentation?
Asymptomatic vs mild mucosal bleeding, petechia, purpura

Labs?
Thrombocytopenia

Treatment?

Platelets > 30K and asymptomatic – conservative/monitor

Platelets < 30K or bleeding; corticosteroids or IV immunoglobulin; splenectomy if refractory

Children w/ ITP typically respond well to steroids

You think they have ITP – What should you test for?

HIV and Hepatitis, maybe cancer screening

47 yo M presents w/ fever, AMS, and lower extremity rash. + IVDU. Petechiae on lower extremities and distal to the BP cuff. Labs show Hgb 8, PLT 20,000, and Cr of 2.8.

Platelets

47 yo M presents w/ fever, AMS, and lower extremity rash. + IVDU. Petechiae on lower extremities and distal to the BP cuff. Labs show Hgb 8, PLT 20,000, and Cr of 2.8.
Thrombotic Thrombocytopenic Purpura (TTP)

Pathophys?

Autoimmune destruction of ADAMTS 13 --> inability to breakdown vWF multimers --> MAHA

Clinical presentation?
Fever, Anemia (MAHA), Thrombocytopenia, Renal failure, Neurologic abnormalities (confusion, seizures, numbness)
(neurologic sx more common in TTP compared to HUS)

Labs?
Anemia, Thrombocytopenia, elevated Cr, Signs of hemolysis (schistocytes, increased LDH, low haptoglobin, increased bili), negative coombs, normal PT/PTT and normal fibrinogen

Treatment?

Plasma exchange transfusion, Steroids, Splenectomy

Platelet transfusion contraindicated

7 y.o. F w/ several days of bloody diarrhea. Other members of the school she attends also had bloody diarrhea. She is irritable and lethargic. Skin appears slightly yellow w/ petechiae to upper extremities. Cr of 3.0 mg/dL, PLT of 38,000, Hgb 8. A peripheral blood smear shows schistocytes.

Even more Platelets

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Hemolytic Uremia Syndrome (HUS)

Pathophys?

Toxin mediated formation of microthrombi that occlude microvasculature. Damaged endothelial cells secrete cytokines that promote vasoconstriction and platelet microthrombus. RBCs destroyed as they pass platelet microthrombi leading to hemolysis, ischemia.

Clinical presentation?

Thrombocytopenia (first), nonimmune microangiopathic hemolytic anemia (second), acute renal failure (hence, "uremia" in title) (third). Sx: Abdominal pain, bloody diarrhea, nausea, vomiting, fatigue, pallor, jaundice

How does this differ from TTP?

Not due to ADAMTS 13 deficiency, usually no neuro deficit

Labs?

CBC: anemia, thrombocytopenia; Elevated serum creatinine; Increased bleeding time ; Normal PT/PTT

Peripheral smear

Schistocytes (helmet cells)

Treatment

Supportive care to promote renal perfusion. No antibiotics.

38 M admitted to the ICU for sepsis develops DVT, petechiae, bleeding at IV sites; labs show anemia, thrombocytopenia, prolonged PT/PTT, low fibrinogen, elevated d-dimer; increased LDH, bilirubin, smear shows schistocytes. Dx?

Heme

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DIC

Pathophys?

Diffuse coagulation resulting in consumption of coagulation factors (consumptive coagulopathy)

Clinical presentation?

Related to an underlying abnormality (severe illness, sepsis, cancer, pregnancy)

Causes include sepsis, trauma, obstetric, transfusion

Mixed picture of bleeding and clotting

Labs?

MAHA, thrombocytopenia, low fibrinogen, elevated d-dimer, prolonged PT/INR & PTT - HIGH YIELD

Treatment

Treat underlying disease and supportive care

65 yo M w easy bruising and gum bleeding. Recently started warfarin for atrial fibrillation. Ecchymoses on his arms and legs. Prolonged prothrombin time (PT) and elevated international normalized ratio (INR). He denies any significant dietary changes. What is the most likely diagnosis, and what is the treatment for this condition?

65 yo M w easy bruising and gum bleeding. Recently started warfarin for atrial fibrillation. Ecchymoses on his arms and legs. Prolonged prothrombin time (PT) and elevated international normalized ratio (INR). He denies any significant dietary changes. What is the most likely diagnosis, and what is the treatment for this condition?
Vitamin K Deficiency (drug induced)

Pathophys?
Deficiency in vitamin K associated factors
Mnemonic: 2 + 7 = 9 not 10 (Factor 2, 7, 9, 10) and protein C, protein S

Clinical presentation?
petechia, bruising, hematomas, oozing at surgical site

Labs?
PT/PTT prolonged
Normal Bleed Time and normal PLTs

High yield facts

Newborns lack gut bacteria that produce vitamin K and need prophylactic vitamin K shot at birth to prevent bleeding

Malabsorptive conditions and long-term antibiotic therapy can cause vitamin K deficiency – (ADEK)

Treatment?
Vitamin K shot, dietary changes, medication change

25 yo F presents with heavy and prolonged menstrual bleeding. She has gone through several extra absorbent tampons with no success. As a child, she had several episodes of unexplained nosebleeds. Labs show normal PT and platelet count with prolonged PTT.

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Von Willebrand Disease

Pathophys?

Deficiency or dysfunction of vWF --> reduced platelet adhesion/impaired primary hemostasis; increased degradation of Factor VIII --> impaired secondary hemostasis.

Clinical presentation

Mucocutaneous bleeding (Easy bruising, Epistaxis, Bleeding of gingiva and gums, Petechiae, Prolonged bleeding from minor injuries), bleeding after surgery, menorrhagia, +FamHx

Classiciagnostic test?

Abnormal Ristocetin cofactor assay

Normal PT/PLTs +/- PTT

Prolonged Bleed time +/- PTT

Treatment

DDAVP(Desmopressin)

3 m.o. infant presents w/ bleeding along the gum line. Petechiae and purpura along the extremities. CBC returns normal platelet count but the infant has increased bleeding time. Dx?

Heme

3 m.o. infant presents w/ bleeding along the gum line. Petechiae and purpura along the extremities. CBC returns normal platelet count but the infant has increased bleeding time. Dx?
Glanzmann's
Pathophys?

AR bleeding disorder caused by decreased GpIIb/IIIa, inhibiting platelet-platelet adhesion/platelet plug formation

Clinical presentation?
Mucocutaneous bleeding from birth, petechiae/purpura

Diagnostic testing?
Normal platelet levels, increased bleeding time

Treatment?
Platelet transfusions

3 m.o. M for persistent bleeding after minor cut. PLT 76,000. Platelet aggregation studies show no response to ristocetin that doesn't correct w/ addition of normal plasma. PBS shows large platelets. Dx?

3 m.o. M for persistent bleeding after minor cut. PLT 76, 000. Platelet aggregation studies show no response to ristocetin that doesn't correct w/ addition of normal plasma. PBS shows large platelets. Dx?

Bernard-Soulier

Pathophys?

AR platelet disorder resulting in a deficiency of GPIb receptor for vWF causing impaired platelet aggregation. Defect in platelet-vessel wall interaction.

Clinical presentation?

Mucocutaneous bleeding

Labs?

Increased bleed time

Abnormal ristocetin assay

BIG SUCKERS

Treatment?

Supportive - avoid anti-platelet medication

Surgery of life-threatening bleed - platelet transfusion

VTE

Risk Factors
Endothelial damage
Venous stasis
Hypercoagulability (cancer, hormones, genetics)
(Virchows triad)

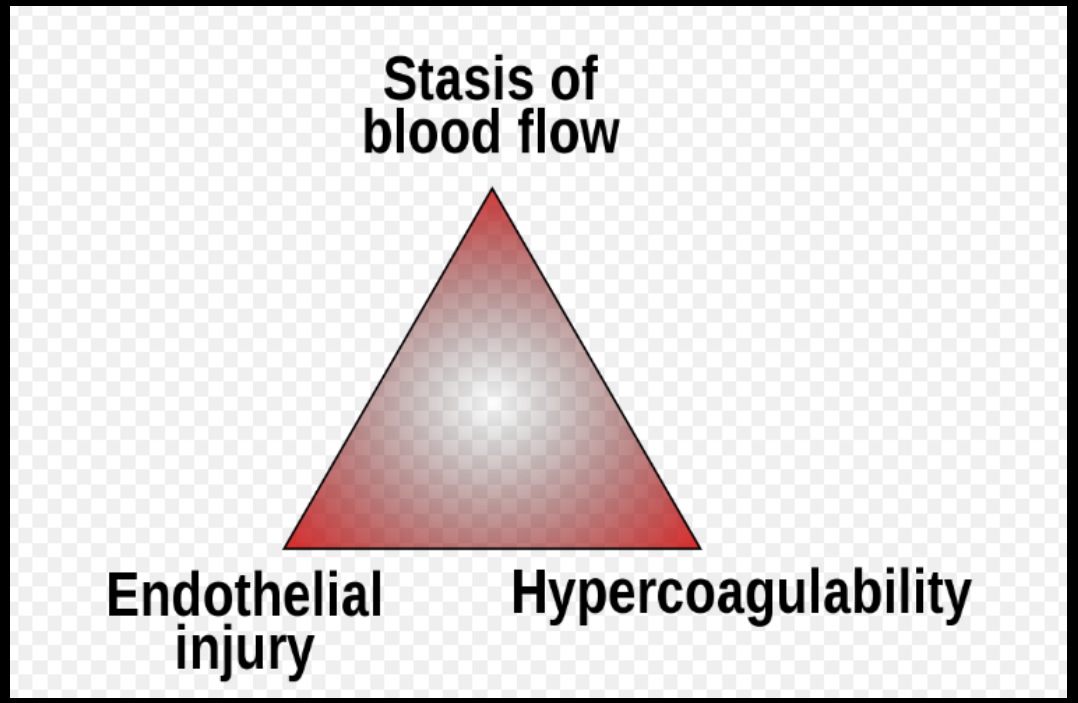
Clinical presentation?
DVT (UNILATERAL warm, swollen, tender lower extremity),
+Homan's sign (not great in real life)
PE (chest pain, dyspnea, fever, tachycardia, tachypnea, hypoxia, death)

Diagnosis?
DVT: Physical exam and lower extremity dopplers
PE: elevated D-dimer; CT angiogram

Treatment?
Fibrinolytic if unstable
Anticoagulants (contrast with antiplatelets)
Prophylaxis typically with LWMH unless kidney disease
Treat with therapeutic heparin, LWMH, DOACs if stable

Most common EKG finding in PE?
Sinus Tachycardia – NOT S1Q3T3

Most common CXR finding in PE?
Nothing



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Approach to Anemia

- Look at the CBC: What's the size of the RBCs -> create DDx from there
  - Microcytic (MCV<80)
  - Normocytic (MCW 80-100)
  - Macrocytic (MCV >100)

65 year old man presenting with 3 month history of fatigue, lightheadedness with exercise, feeling out of breath. On exam you note conjunctival pallor and tachypnea. Next steps?

You obtain CBC which demonstrated hemoglobin of 9 (normal ~15) and MCV 79. You obtain iron studies which show:
Iron 50 (nl 80-180 mcg/dl)
Ferritin 10 (nl 12-300 ng/mL)
TIBC 700 (nl 240-450 mcg/dl)
Transferrin % sat: 10% (nl 20-50%)
Dx?

Microcytic anemia

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Dx?
Iron deficiency anemia

Treatment?
Iron supplementation
What else?
Colonoscopy (new IDA in a 65y.o. is CRC until proven otherwise!), other screening if indicated

Microcytic anemia

TAILS:
Thalassemia (globin chain def)
Anemia of chronic disease
Iron deficiency
Lead poisoning (ALAD)
Sideroblastic anemia (protoporphyrin, ALAS, B6)

Know how to interpret Iron labs:
Serum Iron
TIBC
Ferritin
% sat

CBC:
MCW
RDW

Condition	MCV	Iron	Ferritin	TIBC	Transferrin	Transferrin saturation
Iron deficiency	↓	↓	↓	↑	↑	↓
Inflammatory anaemia	↔	↓	↑	↓	↓	↓
Thalassaemia minor	↓	↔	↔	↔	↔	↔
Thalassaemia major	↓	↔/↑	↑	↓	↓	↑
Sideroblastic anaemia	↓	↑	↑	↔	↔/↑	↑
Iron overload	↔	↑	↑	↓	↓	↑

LearnHaem (https://www.learnhaem.com/courses/anaemia/lessons/iron-deficiency/topic/interpreting-iron-studies/)

55 y.o. F, PMHx significant for RA on MTX, Hashimoto's, and HLD presents for a routine well-visit. She is up to date on all cancer screening, but routine labs show a Hb = 10 with a baseline of 12-13.

Next best step?

55 y.o. F, PMHx significant for RA on MTX, Hashimoto's, and HLD presents for a routine well-visit. She is up to date on all cancer screening, but routine labs show a Hb = 10 with a baseline of 12-13.

Next best step?

Iron studies (still most common)

Not IDA...what is your dx?

ACD

Expected labs? Serum iron, ferritin, TIBC, iron saturation

L, H, L, L

Treatment?

Tx the underlying condition(s)

Pathophysiology?

Inflammation -> increased cytokines/hepcidin (APR) -> less iron transfer from macrophages/less EPO

60 y.o. M with PMHx significant for AUD is being worked up after being found down in the cold. Notably, he was also receiving treatment for TB starting last month. Basic labs show anemia, and iron studies are ordered: ferritin, serum iron, and transferrin are all elevated but TIBC is normal. Dx?

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Sideroblastic anemia

Pathophysiology?

Decreased heme synthesis, iron trapped in mitochondria

Treatment?

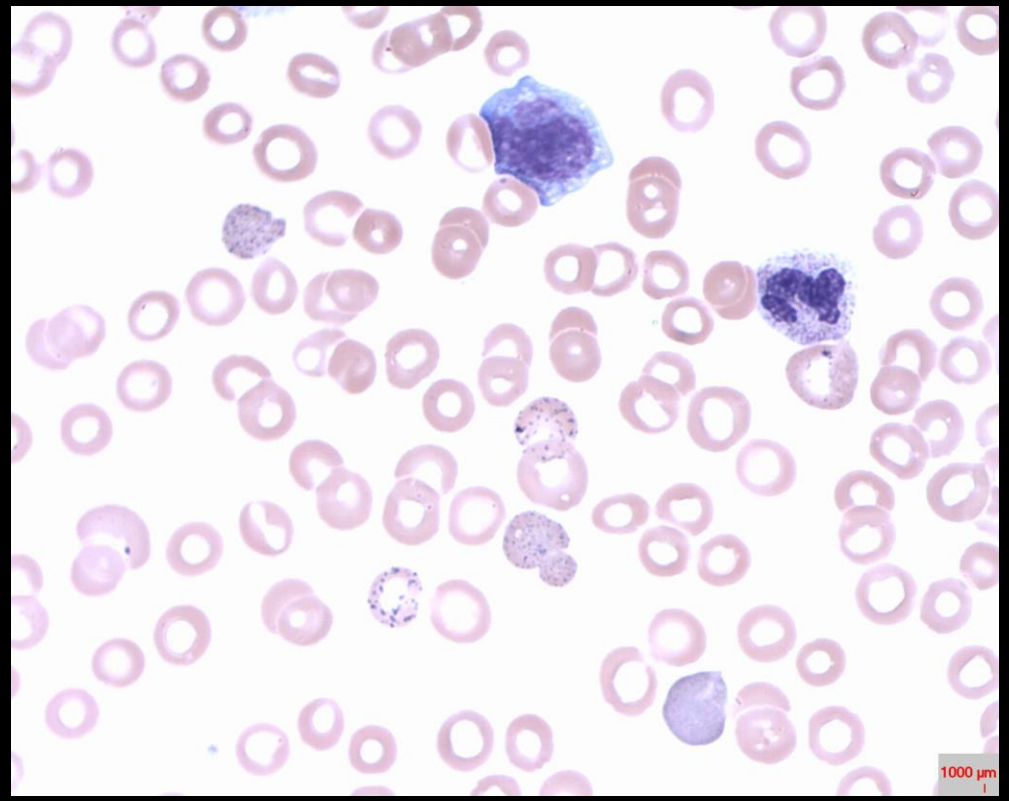
B6 supplementation (cofactor for ALAs)

Cease the offending agent

Causes?

B6 def, INH, Lead, AUD, MDS, Malignancy, Copper def

Sideroblastic anemia



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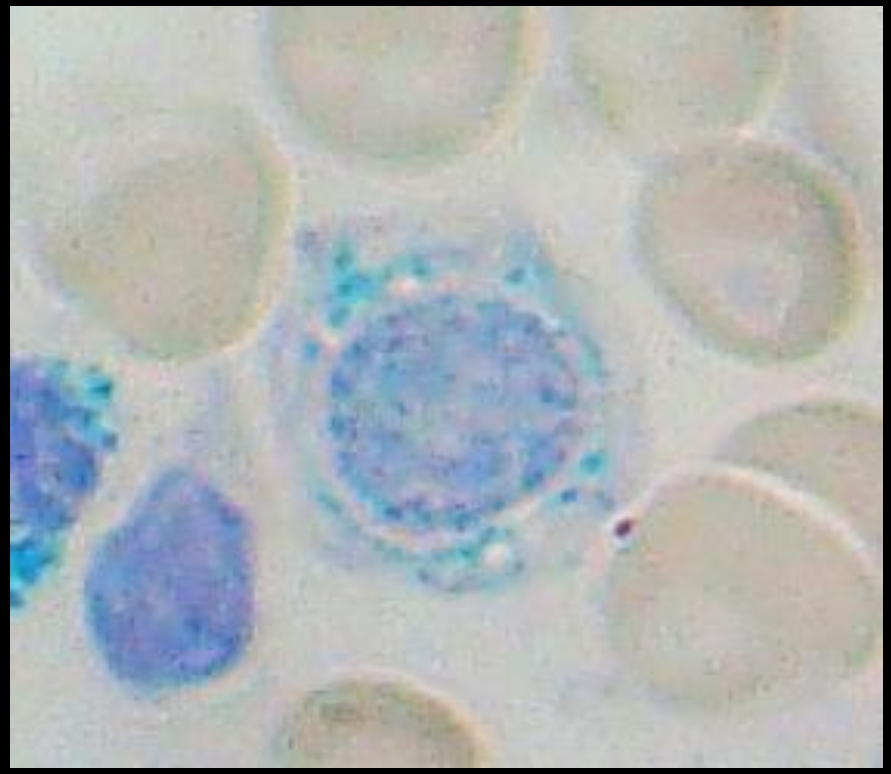
Peripheral smear demonstrating basophilic stippling (sideroblastic anemia, lead poisoning)



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Bone marrow sample with Prussian blue stain demonstrating ringed sideroblasts

Normocytic anemia

MCV 80-100
Consider hemolysis vs nonhemolytic:
How can you differentiate?
Reticulocyte count

Low reticulocyte count/nonhemolytic causes
Early anemia of chronic disease, early iron deficiency – actually HY
Aplastic Anemia
CKD

Hemolytic causes:
Related to the RBC (intrinsic) vs Extrinsic/Extravascular (autoimmune, MAHA, splenic macrophages "hypersplenism")

Intrinisic/intravascular causes include:
Hereditary spherocytosis: elevated MCHC, splenomegaly -> can tx with splenectomy
PNH: defective CD55/59 anchors, complement-mediated hemolysis (DAT negative), tx with eculizumab (anti-C5)
G6PH deficiency: fava beans, TMP-SMX, etc. Heinz bodies/Bite cells on PBS.
Sickle cell disease: pain crises, tx with hydroxyurea + vaccination against encapsulated bugs, daily Penicillin until age 5

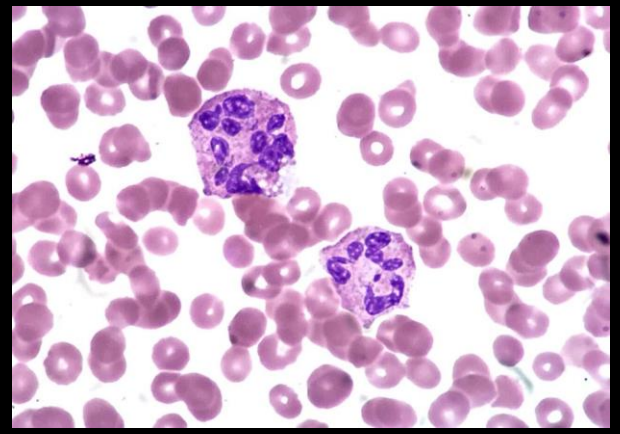
Macrocytic anemia

MCV >100
Consider Megaloblastic vs nonmegaloblastic
How can you differentiate?
Hypersegmented neutrophils

Causes of macrocytic, megaloblastic anemia?
B9 (folate), B12 deficiency

How to differentiate?
MMA – high in B12 deficiency

Ways they test it:
Person with hx of autoimmune disease?
Lean B12 deficiency due to Pernicious anemia
Alcoholic, malnutrition hx?
Lean Folate deficiency...can be difficult
On MTX?
Folate deficiency, supplement folinic acid to prevent



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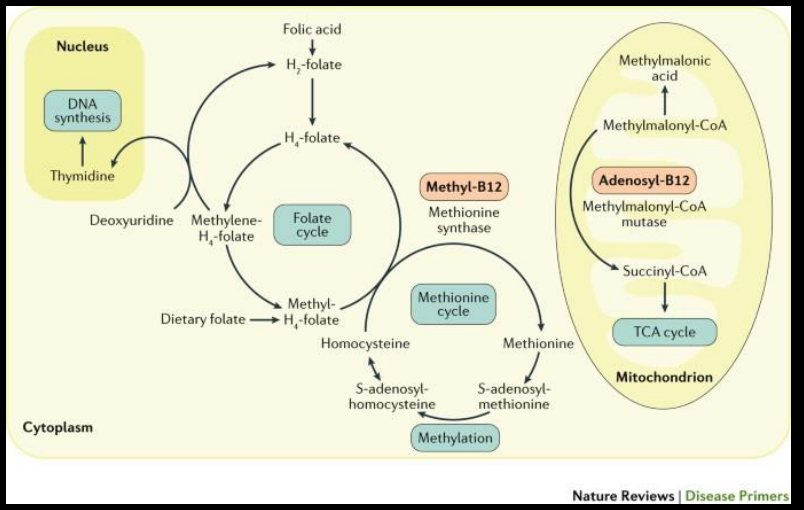

Green, R., Allen, L., Bjørke-Monsen, AL. et al. Vitamin B12 deficiency. Nat Rev Dis Primers 3, 17040 (2017). https://doi.org/10.1038/nrdp.2017.40

42 y/o. F presents to the clinic complaining of diffuse arthralgias and fatigue. She reports no recent infections, and her physical exam is largely benign with exception of a few mucosal ulcers. Initial labs are significant for: Hb 9.6, WBC 1.2, PLT 22k, and a mildly elevated creatinine. Likely diagnosis?

42 y.o. F presents to the clinic complaining of diffuse arthralgias and fatigue. She reports no recent infections, and her physical exam is largely benign with exception of a few mucosal ulcers. Initial labs are significant for: Hb 9.6, WBC 1.2, PLT 22k, and a mildly elevated creatinine. Likely diagnosis?

SLE

Best initial test?

ANA (most sensitive, so good at ruling out if negative)

Most specific test?

anti-dsDNA, anti-Smith

MCC of death in these patients?

Infection, cardiovascular disease

Treatment acutely and for maintenance?

Steroids for flares, hydroxychloroquine for maintenance (watch for retinal toxicity)

What if this patient had a history of recurrent pregnancy loss?

Antiphospholipid syndrome

Treatment?

ASA, ASA + LMWH in pregnancy

ID cases to keep in mind…

Cyclical fever with hemolytic labs?

Malaria, Babesiosis

Travel to the carribean, diffuse myalgias/bone pain, thrombocytopenia?

Dengue hemorrhagic fever (Chikingunya also tested)

Kid with SCD gets profoundly anemic with no retic count?

Parvovirus B19 infection (aplastic crisis)

High-school kid with dry cough, atypical PNA has a hemolytic anemia?

Mycoplasma, cold-agglutinin disease

Pharm cases to keep in mind...

35 y.o. F at 12 weeks pregnancy, found to have Grave's disease. Now has leukopenia. Cause?

PTU/Methimazole

30 y.o. M with hx of treatment-refractory Schizophrenia is found to have PNA. CBC shows leukopenia. Cause?

Clozapine

25 y.o. M with hx of IBD is found to be leukopenic. Cause?

Sulfasalazine

40 y.o. F with hx of refractory epilepsy is found to be hyponatremic and leukopenic. Cause?

Carbamazepine

A 65 y.o. M with PMHx of CAD s/p PCI x2, PAD, HLD, and T2DM presents to an OSH with crushing chest pain. An EKG is similar to prior exams, and his troponins are mildly elevated. Prior to transfer to a tertiary center, he is started on heparin, a beta-blocker, and a nitroglycerin drip that results in improvement of his chest pain. Upon transfer, his current regimen is continued, and his cath gets delayed multiple times due to emergent STEMIs. He is then noted by his nurses to be cyanotic and hypoxic to 85% despite 4L NC O2, but he only complains of a headache. Likely dx?

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Methemoglobinemia

Best next step?

ABG/co-oximetry

Treatment?

Stop the nitro, methylene blue if symptomatic (confusion, etc.)/level >30%

Classic exam finding?

Chocolate-colored blood

40 y.o. F presents to the ED with intractable headache. PMHx is significant for migraine, but this episode feels different. Recent history is notable for recently being at a birthday party with an indoor grill. Next best step?

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ABG with co-oximetry

What if there was a house fire with inhalation injury on exam? Next best step?

Intubate!, also consider giving cyanocobalamin (associated cyanide tox.)

Treatment?

100% O2 if mild – what else?

HYPERBARIC O2 – HY!

Neurotoxicity sx (severe HA) should get an MRI – what do you see?

b/l globus pallidus involvement

What would you expect the ABG to look like: PaO2, PaCO2, pH?

NORMAL PaO2, probably acidotic

52 y.o. M presents to the clinic with the complaint of blurry vision. He has also noted a darkened skin tone on his face, but it's summer. He also describes episodes of intense itching that are worse when he uses the hottub at his gym. Vitals are all wnl. Next best step?

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Check a CBC (Hb/Hct) - if elevated without hypoxia = suspicious

Confirmatory test?

Genetic testing – JAK2 mutation

Expected EPO level?

LOW (would be high in secondary polycythemia 2/2 hypoxia)

Treatment?

Phlebotomy + ASA +/- hydroxyurea

Most tested complication?

Thrombosis – portal vein thrombosis, stroke, etc.

A 25 y.o. M presents to the clinic after an odd episode of pain in his neck associated with drinking alcohol. He reports 2 month history of night sweats and a swelling in his neck. On exam, there is shotty cervical LAD with one dominant node measuring ~5cm.

Next best step?

A 25 y.o. M presents to the clinic after an odd episode of pain in his neck associated with drinking alcohol. He reports 2 month history of night sweats and a swelling in his neck. On exam, there is shotty cervical LAD with one dominant node measuring ~5cm.

Next best step?

Lymph node biopsy – findings?

Then what?

CT/PET-CT for staging (actually HY)

They decide to treat with ABVD (doxo, bleo, vinblastine, dacarbazine). What tests should be done?

Echo (for doxo), PFTs (for bleo)

They also decide to do radiation to the chest for mediastinal LAD seen on CT...complications?

Constrictive pericarditis, secondary malignancy (very HY)

80 y.o. M presents to the ED with s/sx of PNA. Per the patient, he has also noted multiple swollen glands in his neck and axillae. Chart review reveals 20lbs of weight loss in the past year. He reports feeling extra fatigued lately. On exam, his spleen is palpable. Labs are largely unremarkable. Next best step in dx?

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CLL-Flow cytometry (expect CD5, 19, 20)

You get a smear – what would you see?

Smudge cells

Treatment?

Observation vs targeted therapy if super symptomatic

50 y.o. F presents with diffuse lymphadenopathy, unintentional weight loss, and progressive fatigue. A CBC is done and are significant for WBC 35k with eosinophilia, Hb 9.6, and PLT 250k. Likely Dx?

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CML

Confirmatory steps?

Bone marrow, cytogenetics showing t9:22

Expected LAP level?

Low (high in leukemoid rxns)

Treatment?

Imatinib (tyrosine kinase inhibitor)

A 60 y.o. M with recently diagnosed AML undergoes induction therapy. After his treatment, he is monitored. He begins to be lethargic, with associated muscle cramping and runs of VT on telemetry. Dx?

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Tumor Lysis Syndrome

Labs?

Hyperkalemia, hyperphosphatemia, hypocalcemia, hyperuricemia

Prevention?

IV fluids (most important) +/- allopurinol

Patient is also noted to have an AKI. Why?

Crystalline nephropathy 2/2 uric acid causing tubular obstruction

A 65 y.o. M presents to the ED with a 5-day history of worsening fatigue and malase with associated dark urine. Initial labs are significant for: WBC 56k, PLT 30k, Hb 7.2, elevated LDH, low haptoglobin, and elevated PT/aPTT. A PBS is done and shows blasts with MPO-positive, needle-shaped intracellular inclusions. Most likely diagnosis?

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APML (subtype of AML)

PBS finding?

Auer Rods

Translocation?

t15:17

Treatment?

ATRA – leads to differentiation of blast cells

A 64 y.o. M with PMHx of CAD s/p PCI is transported to the ED after an MVC where he sustained several injuries. Due to hypotension and initial Hb 7.2 with signs of active external bleeding, he is transfused with massive transfusion protocol (1:1:1 ratio) with 3u pRBC. His bleeding stabilizes, and vitals have stabilized within 6 hours of arrival. However, he then notices leg swelling and shortness of breath requiring up to 4L O2 via NC. A STAT CXR shows signs of pulmonary venous congestion and diffuse infiltrates. Dx?

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TACO (Transfusion-Associated Circulatory Overload)

Pathophysiology?

Fluid overload due to large-volume transfusions in susceptible pts

Tx?

Diuretics (tx like a HF exacerbation)

A 55 y.o. F with a hx of alcholic cirrhosis and COPD presents to the ED after being found to be anemic to Hb 8.9 at her PCP. This is an acute change over the last few days, and the patient reports dark stool with associated fatigue and occasional shortness of breath. GI is consulted for suspected upper GI bleed, and FFP is transfused in the setting of coagulopathy due to underlying liver disease. 2 hours later, the patient is hypoxic, tachypneic, and hypotensive. A STAT CXR shows diffuse interstitial infiltration. Most likely diagnosis?

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TRALI (Transfusion-Associated Lung Injury)

PPx?

Primed neutrophils in the lung endothelium being activated by substances in the transfusion = NONCARDIOGENIC pulmonary edema

Tx?

Aggressive supportive care (respiratory support, fluids, pressors, etc.)

A 72 y.o. M presents to the clinic with a CC of back pain. He reports this has been progressively worsening over the recent months. He has also struggled with consitpation during this time. On exam, you feel that he has midline spinal tenderness to palpation, and he has some mild-moderate peripheral edema. Initial labs are significant for: Ca 11.7, Hb 9.7, and Creatinine 1.6 with baseline ~0.9. Likely dx?

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Multiple Myeloma

Typical sx?

CRAB (hypercalcemia, renal disease, anemia, and lytic bone lesions)

Expected findings on UA

Proteinuria – nephrotic syndrome (>3.5g/d)

Expected findings on blood smear?

Rouleaux formation

Best initial diagnostic test?

SPEP – looking for "M-spike"

Confirmatory test?

Bone marrow biopsy showing clusters of plasma cells

Leading cause of death?

Infection – antibodies you make are not very funtional

Thank you for watching!

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